Study of Pulmonary Hypertension in Egyptian children and adolescents with β-Thalassemia Intermedia

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Background:

Pulmonary hypertension (PHT) and increased thrombosis are the commonest pattern of cardiovascular involvement among adult patients with â-thalassemia intermedia (TI). In Egypt, beta-thalassemia is the most common chronic hemolytic anemia (85.1%). A carrier rate of 9-10.2% has been estimated in 1000 normal random subjects from different geographical areas of Egypt.

Objective:

The purpose of our study was to detect the prevalence of PHT among TI patients, its relation to clinical and laboratory characteristics and possible effects of therapeutic strategies used in management those patients.

Materials and Methods:

In this study,60 consecutive â-TI patients followed up in Pediatric Hematology Clinic, New Children's hospital, Cairo University, (24 males and 36 females), aged 6-28 years with mean age 13.5±6.6 years were evaluated by trans-thoracic two dimensional (2D) guided (M mode) and Doppler echocardiogram; mean pulmonary artery pressure (PAP) was calculated using the formula: mean PAP = 0.65 PASP + 0.55 mmHg. PHT was diagnosed if mean PAP was > 25 mmHg. Myocardial performance index (MPI) was calculated as the ratio of the sum of iso- volumetric contraction and relaxation times over the ejection time We further tested for possible risk factors, through comparing 32 patients with evidence of PHT (Group I) to 28 age- and sex-matched TI patients without PHT (Group II).

Results:

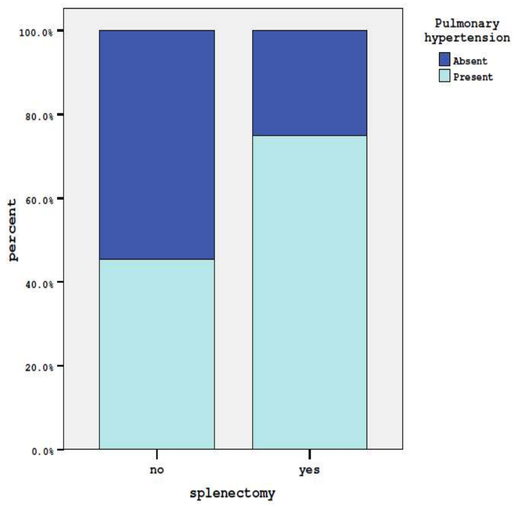
The overall prevalence of PHT was 53.3% (48.9% in pediatric patients and 66.7% in adults).

Table 1: Comparison of Baseline Data of group I & group II Patients:

Variables	Group I (n=32)	Group II (n=28)	P-value
	Mean±SD	Mean±SD	
Platelets(x10^3/ ccm:	498.4±296.7	331±131	0.048*
HbF (%)	29.9±23.6	27.5±21.4	0.9
Serum ferritin (ng/ml):	959.3±686	642±618.9	0.024*
Mean Follow up duration (yrs)	9.8±6.4 (8.5; 0.5-12)	6.3±5.0 (4.0; 1-21)	0.03*
Duration of L-Carnitine(yrs)	5.4±4 (4; 0.33 – 19)	3±2 (3; 0.33 – 8)	0.02*
Splenectomy No.(%)	12 (37.5%)	4 (14.2%)	0.04*
Transfused No.(%)	28(53.8%)	24(46.2%)	1.00
Chelation intake No(%)	15 (46.9%)	6 (21.4%)	0.04*

As presented in table (1) Splenectomy (p=0.04), long duration of follow-up (p=0.03), high mean platelet counts (p<0.05), and high serum ferritin (p=0.02) were associated with PHT.

Figure (1) shows that splenectomized patients have statistically significant higher incidence of PHT



There was a weak proportional correlation between mean PAP and the duration of follow-up (r=0.31, p=0.02), reticulocytic count (r=0.3, p=0.04), total bilirubin (r=0.3, p=0.04) and serum ferritin (r=0.3, p=0.03). Myocardial performance index of right ventricle (MPI of RV) was proportionally correlated to mean PAP.

Logistic regression analysis performed revealed that mean platelet count was the only independent variable which retained its statistical significance (OR 1.004 (95% CI: 1.0-1.008, p=0.045).

Variable	r-coefficient	P value
Duration of follow up (years)	0.31	0.02*
Splenectomydurati on(years)	-0.13	0.63
Hb (g/dl)	-0.25	0.059
Platelets (x103/ccm)	0.24	0.07
Reticulocytic count (%)	0.27	0.039*
Total bilirubin (mg/dl)	0.27	0.037*
LDH (u/l)	-0.06	0.66
Serum Ferritin (ng/ml)	0.28	0.03*
MPI (RV)	0.4	0.001*

Conclusion: Our results support recent studies reporting a high prevalence of PHT . Possible risk factors include long duration of disease, splenectomy, high serum ferritin, high platelet count and increased hemolytic rate. Early screening of splenectomized patients with thrombocytosis is highly recommended. Further prospective long- term studies evaluating the effect of hydroxyurea and chelation therapy on PHT are required.

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